CHEMODECTOMA OF THE HEAD AND NECK

OF GLONUS TUMORS OF THE EAR

ESSAY

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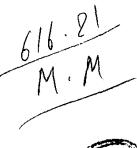
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INTRODUCTION

Chemodectomas or non-chromaffin paragangliomas are a histologically similar group of tumors that arise from chemoreceptive tissues throughout the body. These tumors are commonly found in the head and neck as carotid body tumor, glomus jugulare tumars, glomus vagale tumors, or glomus tympanicus tumors.

These tumors are indistinguishable by microscopic examination. The classification of chemodectomas can be made according to the anatomic site of the chemoreceptor tissue from which the tumor originated. The clinical appearance, diagnosis and management of chemodectomas vary with their sites of origin.

HISTORICAL REVIEW

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Von Haller was the first to describe the carotid body in 1743. The first carotid body tumor was described by Marchand in 1891.

According to Guild (1941), the name glomus jugulare was given to small structures in human temporal bone sections located near or in the wall of the jugular bulb.

Rosen Wasser (1942), was the first to recognize the clinical entity of the glomus tumor and its origin from the glomus bodies.

In 1949, Lattes and Waltner gave the name non-chromaffin paragangliomas because this name more accurately indicates their cellular origin.

According to Mulligan (1950), and Glenner and Grimley (1974), the name chemodectoma was given since these tumors arise from chemoreceptor cells.

According to Glasscock el al (1974), Glomus tumor is the most commonly used and accepted term. There is tendency to refer to those lesions involving the middle ear and mastoid only as glomus tympanicum and those primarily in the bulb as

glomus jugulare tumors.

Muratori in 1932 was the first to describe nests of chemoreceptor cells in the ganglion modosum of birds.

Similar aggregates of cells were found in the perineurium of the vagus nerve of man below the ganglion nodosum. Stout in 1935 reported the first case of a tumor of the nodose ganglion of the vagus nerve. The term "vagal body tumor" was given by Birrell in 1953 to designate the chemodectoma that arises from the chemoreceptor tissue of the vagal body.

Watzka (1963), was the first to describe non-chromaffin paraganglia in the plica ventricularis of the larynx. He described a paired superior laryngeal glomus body lying in the supraglottic region just above the anterior ends of the vocal cords, in relation to the internal branch of the superior laryngeal nerve. It is called glomus laryngicum superior. The other pair was described by Kleinsasser (1964), the glomus Laryngicum inferior which lies in the subglottic region in relation to the recurrent laryngeal nerve. Kleinsasser 1964, also found a further paraganglia on the anterior aspect of the larynx which he called the glomus laryngicum anterior.

According to Lawson and Zak (1974), minute inconstant nests of aberrant paraganglicaic tissues may be found anterior to the cricoid cartilage, lateral to it, posteriorly in relation to the transverse inter-arytenoid muscle or in the capsule of the thyroid gland, these may be considered to be ectopic inferior laryngeal paraganglionic tissue.

According to Schuller and Lucas (1982), paraganglioma can occur in nasopharynx. This is extremely rare.

CLASSIFICATION OF PARAGANGLIOMAS

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The paraganglia located outside of the adrenal medulla are analogous to the adrenal medulla paraganglia but Lack the batter's affinity for chromium salts, and therefore tumors arising from these are referred to as non-chromaffin paragangliomas (Similarly, tumors of the adrenal medulla, though commonly referred to as pheochromocytomas, are also referred to as chromaffin paragangliomas).

A few of the nonchromaffin paragangliomas have chemoreceptor activity and are referred to as chemodectomas

(carotid and aortic body tumors would be included in this
category).

This complex terminology may be replaced by an anatomic classification recently proposed by Glenner and Grimky (1974), in which extra-adrenal paragangliomas are subdivided by location into the following categories:-

- branchiomeric (carotid, jugular, laryngeal, subclavian, aortic and pulmonary).
- 2. Intravagal (upper mediastinum).
- 3. Aorticosympathetic (retroperitoneal).
- 4. Visceral autonomic (other viscera).

EMBRYOLOGY - ANATOMY - PHYSIOLOGY
OF NON-CHROMATTIN PARAGANGLIA

EMBRYOLOGY

The chemorecepter cells are derived from the neuroepithelial elements of cranial nerves that migrate to localized areas of mesodermal condensation. These areas are
situated around vessels of embryonic branchial arches
(Zachariah, and Shah, 1972).

According to Rosen et al (1981), the carotid and glomus bodies are derived from meural crest paraganglion cells.

According to Maran (1979), the neural crest differentiates into the Schwann cell and the sympathicoblast; this latter cell gives rise to paraganglionic cells from which arise chemodectomas and glomus jugulare tumors, and ganglionic cells from which arise benign and malignant ganglioms.

ANATOMY

The paraganglia are divided into :-

- a. Sympathogenic chromaffin paraganglia.
- b. Non-sympathogenic non-chromaffin paraganglia.

The chromaffin paraganglia include the medulla of the suprarenal gland and various smaller paraganglia in the body.

There are also individual scattered chromaffin cells or groups of cells in the branches and neurones of the sympathetic nerve.

The chromaffin paraganglionic system is part of the sympathetic system.

The non-chromaffin paraganglia belong to the parasympa thetic system and may lie in the adventitia of blood vessels and along the vagus and the glossopharyngeal nerve (Fig. 1).

The most important non-chromaffin paraganglia are :-

- 1. Glomus caroticum (carotid body).
- 2. Glomus jugulare.
- 3. Glomus tympanicum.
- 4. Glomus juxta-vagale and Glomus intravagale.
- 5. Glomus laryngicum (superior § inferior).

Glomus Caroticum (Carotid body);~

The carotid body occurs bilaterally and is located at or near the bifurcation of the common carotid artery. It is attached to the bifurcation by a firm connective tissue band. It is usually evoid in shape, averages 6 by 4 by 2 mm. in size and varies in colour from brownish red to gray. It possesses a fibrous capsule with fibrous septa. The blood supply is derived from a small artery arising from the internal, external or common carotid arteries. The smaller vessels reach the parenchyma by means of the dividing fibrous septa which usually show marked vascularity.

The carotid body is richly supplied by nerve fibers of sympathetic and parasympathetic origin. The sympathetic fibers are derived from the cervical sympathetic ganglia and the parasympathetic fibers accompany the vagus, hypoglossal and glossopharyngeal nerves. Like all organs of the body the carotid body becomes small and atrophic with the advance of age.

Histologically, the carotid body consists of fibrous stroma and parenchyma. The fibrous stroma consists of a capsule